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CASE REPORTS ON FIBROUS DYSPLASIA OF ORBIT

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Abstract : Fibrous dysplasia is a non neoplastic, hamartomatous, developmental disease of bone of unknown aetiology.It is most commonly seen in the maxilla and mandible in the facial skeleton.Involvement of zygoma is rare.We present 4 cases of fibrous dysplasia with one patient having isolated zygomatic bone involvement which is a rare presentation.

Keyword :fibrous dysplasia, hamartoma, zygoma.

CASE REPORT

An 8yr old female child presented with complaints of swelling beneath the left lower lid since birth and watering left eye for 6 months. The swelling was not progressive. On examination, there was a diffuse swelling in the left lower lid extending to the maxilla. The swelling was firm, non-tender and ill-defined (fig.1). Ocular examination was found to be normal with full EOM and 6/6 vision on snellen's chart.

Fig.1 (Case 1) clinical photograph showing diffuse left lower lid swelling

CT imaging was done which showed hyperdense and hypodense lesions representing osteosclerotic and osteolytic areas respectively in the zygomatic region (fig.2, 3).

Fig.2 CT ORBIT coronal view showing lesion involving zygoma of case 1

Fig.3 CT ORBIT axial view of case 1

FNAC was done which showed benign spindle cells with fibrous tissue in the trabeculae of immature woven bone suggestive of fibrous dysplasia (fig.4).

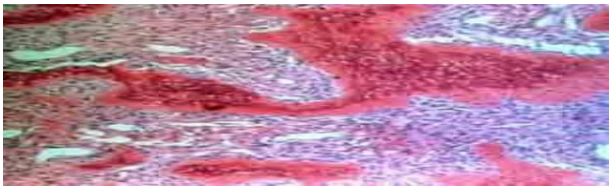

Fig.4 microphotograph showing benign spindle cells with fibrous tissue in trabeculae of immature woven bone

The second patient was a 38yr old female who presented with complaints of progressive protrusion of right eye since childhood associated with pain on and off. On examination, there was eccentric proptosis (fig.5) with horizontal displacement of globe by 1.5mm.There was fullness in the medial aspect without any palpable mass

Fig 5 (Case 2) clinical photograph showing eccentric proptosis

CT imaging showed expansile lesion involving right frontal, ethmoid, sphenoid sinus and nasal septum and partially involving maxillary sinus. Right optic foramen encroachment was also presented suggestive of fibrous dysplasia (fig.6). Biopsy also confirmed the diagnosis.

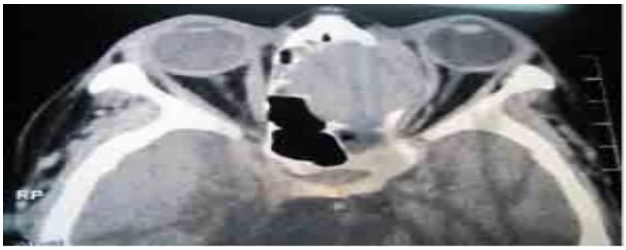

Fig.6 CT orbit axial view showing expansile lesion in case 2

The third patient was a 23 year old male who presented with downward displacement of left eye (fig.7). Ocular examination of left eye was normal with 6/6 vision and full EOM, bony hard swelling was palpated in supero lateral orbit continuous with superior orbital margin and fundus showed choroidal folds near macula. CT imaging showed bony thickening of left frontal bone (fig.8).

Fig.7 Clinical photograph of case 3

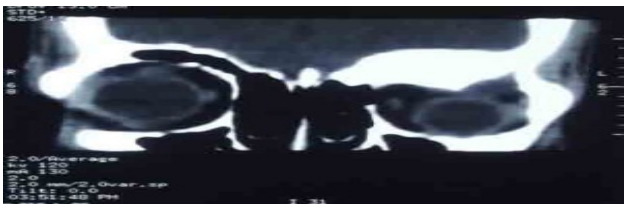

Fig.8 CT coronal view showing hyperostosis of frontal bone

The fourth patient was a 26 year old female who presented with defective vision and protrusion of left eye. Bony hard swelling was palpated in the superolateral orbit continuous with the orbital margin (fig.9). Ocular examination showed restriction of abduction with 6/24 vision and fundus showing choroidal folds. CT orbit showed osteosclerotic bony lesion involving the left frontal bone(fig.10).

Fig.9 Clinical photograph of case 4 showing eccentric proptosis

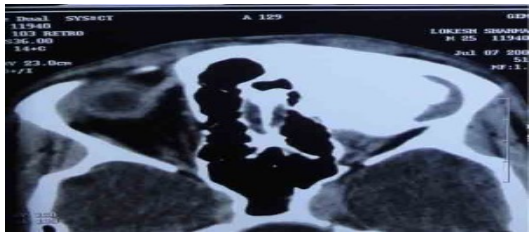

Fig.10 CT axial view showing hyper ostotic lesion in frontal bone

DISCUSSION:

Fibrous dysplasia is a nonheritable pathological condition in which normal bone is replaced by abnormal fibrous tissue. It is considered to be due to arrest of bone maturation at woven bone stage[2]. Lichtenstein first coined the term in 1938. It constitutes 2.5% of all bony neoplasms and 7% of benign bony lesions. Common sites involved are femur, tibia, fibula, ribs and facial bones. Maxilla is commonly involved in the cranio-facial region. Zygomatic bone involvement is very rare. There is no sex predilection though female preponderance has been seen in some studies. Age at presentation is usually first or second decade[1]. There are 2 types of fibrous dysplasia with mono-ostotic form accounting for 80% of cases and poly-ostotic form in 20% of cases.10% of mono-ostotic and 50% of poly-ostotic lesions involve craniofacial bones. Poly-ostotic variety with cutaneous pigmentation and precocious puberty is called Albright syndrome[3].

Pathophysiology:

Fibrous dysplasia is caused by the sporadic mutation of the *GNAS1* gene, which encodes the alpha subunit of the stimulatory G protein (G1), located on chromosome 20q13.2-13.3 of the osteoblastic cells[4]. There is a substitution of cysteine or histidine, amino acids of the genomic DNA in the osteoblastic cells, by another amino acid, arginine. Osteoblastic cells expressing this mutation have a higher DNA synthesis than normal bone cells. The growth of these cells is faster, leading to an inappropriate differentiation of mesenchymal cells[4].

Fibrous dysplasia is a slow growing lesion. Symptoms depend on site of involvement. Involvement of zygomatic bone may cause swelling, facial asymmetry, pain, diplopia, proptosis and loss of vision. Nasolacrimal duct blockage, diplopia, nasal obstruction, malocclusion, raised intra cranial pressure and cranial nerve palsies also occur. Acute or subacute compressive optic neuropathy can arise due to intralesional hemorrhage,sphenoidal mucocele, or secondary aneurysmal bone cyst. Less than 1% of cases transform into malignancy[5].

CT is useful in the diagnosis by showing ground glass appearance of involved bone. Fibrous dysplasia of the cranio facial bones had a pagetoid pattern (56%) to be the most common pattern followed by sclerotic (23%) and cyst like changes (21%). MRI helps in differentiating it from hyperostotic meningioma, aneurysmal bone cyst and giant cell tumour[6]. Malignant transformation in to osteosarcoma, fibrosarcoma,
chondrosarcoma and giant cell sarcoma occurs in 0.4%-0.5% of cases.

Treatment is not required in all patients. Indications for surgical intervention are pain, sarcomatous transformation, cosmesis, functional deficit and for prevention of optic nerve compression. Surgical excision for well localised lesion or curettage with bone grafting can be done[1].

Conclusion:

We have presented 4 cases of fibrous dysplasia one being mono-ostotic variety involving the zygoma which is uncommon. Though the condition presents before the 3 rd decade, it can also be seen in older individuals being asymptomatic for a long duration which is the case in one of our patients.

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